MASTERCLASS *Clinical cases from the Royal National Hospital for Rheumatic Diseases*

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Diagnosis and management of algodystrophy

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The syndrome of pain and swelling occurring after trauma or burn has been recognised for many centuries, but it was Weir Mitchell who first reported the classical features of the syndrome during the American Civil War which he termed 'causalgia'.¹ Since that time many terms have been used to describe this syndrome, including 'algodystrophy', 'algoneurodystrophy', 'reflex sympathetic dystrophy', and 'Sudeck's atrophy' among others, to emphasise the presumed underlying neurological disturbance.²⁻³ In general, the condition is characterised by pain, vasomotor disturbance, and trophic changes affecting part or the whole of the limb. Other sites, particularly the spine, may occasionally be affected.³⁻⁴ Algodystrophy may also occur after minor soft tissue injury or, as in up to half the cases, no injury at all. Sudeck described the post-traumatic severe syndrome with marked bony changes.⁵ This condition and bipolar shoulder/hand syndrome⁶ are the most commonly recognised forms.

Case history

A 56 year old woman, who in December 1982 slipped on a floor, sustained a trimalleolar fracture of the right ankle. Surgical reduction was achieved and the plaster was removed in March 1983. Within 20 days of this she presented with a cold purple foot and had radiographic patchy osteopenic changes. The diagnosis of algodystrophy was made. Failure to improve resulted in the removal of stabilising pins, followed by manipulation under anaesthesia, but postoperatively the foot became stiff. She therefore underwent chemical sympathectomies followed by surgical sympathectomy, but all these measures failed so that ultimately she had an elective below knee amputation in June 1986. The immediate postoperative period was complicated by contractures of the muscles and atrophy, leaving a prominent and tender bony tubercle, which eventually became pain free. Fortunately, she did not develop a phantom limb syndrome.

In August 1988 she fell on an outstretched left hand and sustained an undisplaced Colles' fracture of the left wrist. She was placed in a plaster slab, which was removed fairly early in view of her past problems. Unfortunately, despite this the wrist became stiff with flexion deformities of the proximal interphalangeal joints of the ring and little fingers. In addition, from the forearm distally the arm was cold and

purple with shiny skin. Despite physiotherapy and a series of regional guanethidine blocks and nifedipine she had severe and continuous pain with reduced function. Contractures became progressively worse, and she was referred to the Royal National Hospital for Rheumatic Diseases.

On arrival here it was noted that the left upper extremity from the mid-forearm distally was red, warm, dry, and diffusely tender. The skin was shiny and taut and there were flexion contractures of all the fingers, absent finger movements, wasting of the muscles, and markedly reduced hand function. Movements of the shoulders were normal and there were no other abnormalities except for the below knee amputation on the right. Radiographs of the right hand showed diffuse osteopenia, and thermography demonstrated a hot left hand and wrist. Results of routine biochemical and haematological tests were normal.

She was treated with intensive physiotherapy and salmon calcitonin 100 units daily for 21 days. Within days the pain decreased and hand function slowly improved. At the end of the treatment hand function had returned to 70% of normal, though there were still flexion deformities of the ring and little fingers, which were painful after use. Apart from this, pain was minimal and there were no vasomotor changes. She was discharged on a regular exercise programme.

Discussion

Diagnosis of algodystrophy is essentially clinical. Pain severity (often burning) out of proportion to the preceding injury and its persistence is the clue. Other typical features include hyperaesthesiae, vasomotor changes, hyperhydrosis, and trophic changes. It occurs at all ages, in women more than men, and the incidence increases until late middle age. Pregnancy may be a predisposing cause.⁷ In the fully developed case diagnosis is fairly easy, but partial forms are common and in many pain predominates. Classical stages of early hyperaemia and swelling, subsequent ischaemic dystrophy, and eventually an atrophic state vary greatly in time scale. Early changes are often not seen and progression to dystrophy/atrophy may not occur. In addition, vasomotor changes leading to skin cooling and mottling may be intermittent. Although it is clinically difficult to define such a variable polymorphic condition, it may be

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Table 1 Clinical grades of algodystrophy

	Grade 1 (severe)	Grade 2 (moderate)	Grade 3 (mild)
Pain	Stabbing, lancinating, burning	Dull, aching, diffuse	Insidious onset
Vasomotor	Severe vasoconstriction (or vasodilatation)	Skin cold and clammy or hot and dry	Mild but different from unaffected side
Trophic	Fixed. Atrophy of skin, subcutaneous tissue, muscles and bone	With or without osteoporosis on x ray	Absent, with or without oedema

Table 2 Clinical stages of algodystrophy

	Stage 1	Stage 2	Stage 3
Onset after injury/duration	Days to weeks	Months	Months to years
Vasomotor	Vasodilatation	Vasoconstriction	Vasoconstriction
Skin/trophic changes	Erythema, sweating, oedema	Pallor, cyanosis sweating, indurated swelling, patchy osteoporosis	Pallor, cyanosis, dry, thin skin. Tissue atrophy. Diffuse osteoporosis

considered in terms of either gradation (table 1) or stage (table 2) of disease.

Hand and foot involvement is well recognised and this may spread proximally, but primary knee involvement has only recently been well reported.⁸ As pain and vasomotor changes may be the only obvious manifestations it may in fact be a common cause of chronic knee pain.⁹ A whole limb may become affected or the disorder may be confined to a digit.¹⁰

Pathogenesis is poorly understood. It is believed that sensitised wide range multireceptive neurones in the spinal internuncial neurone pool are at the centre of an abnormal reflex, resulting in excessive sympathetic outflow. For unknown reasons sensitisation occurs after initial nociceptive afferent stimulation, which subsequently results in abnormal pain perception and increased sympathetic afferent activity. Persistent afferent input from damaged soft tissue nociceptive fibres was thought to be important. This input is now known not to continue once a tissue has healed and yet many cases of algodystrophy occur without dystrophic changes. Instead it is postulated that, subsequently, abnormal response to mechanoreceptor afferents occurs (figure).¹¹ Expansion of the area of sensitised wide range multireceptive

neurons within the spinal cord can then give rise to spread of the condition. The influence of descending pathways explains an association with cerebral lesions. Psychological status might affect the condition by this mechanism. Algodystrophy is not a hysterical disuse state, but a withdrawn, passive, and sometimes depressive affect is not infrequently found in patients. A subgroup probably does have a psychiatric disorder as a factor in both the precipitation and perpetuation of the condition, though cause and effect are difficult to distinguish.¹²

Alternative local tissue mechanisms have recently been proposed as being important, whereby a series of vicious circles is established, in which vasodilatation, low flow, and persistent stimulation of nociceptors all play a part.¹³ The authors also suggest that alteration in adrenoceptors may be involved. A possible role for oxygen free radicals has been postulated after a short term study of dimethyl sulphoxide as a treatment for algodystrophy.¹⁴ Four cases have been reported as having apparently occurred after borrelia infection.¹⁵ Deposits of IgG have been found in the palmar fascia of several patients developing an atypical form of algodystrophy in association with neoplasia at another site, suggesting a possible immunological factor, at least in these cases.¹⁶

As abnormal levels of pain are central to the condition, assessment of its severity is important, especially in monitoring patient progress, but is difficult. Multiple measures of pain severity have been advocated,¹⁷ though a simple visual analogue scale is generally the most useful, especially when used in conjunction with a joint tenderness score. The use of a dolorimeter may give a more reliable and objective assessment of tenderness.¹⁸

Infrared thermography is a useful non-invasive method by which vasomotor abnormality of the affected limb can be assessed objectively. Whether the extension of the technique using a mild cold stress test for upper limb algodystrophy increases diagnostic and severity assessment sensitivity is uncertain.¹⁹ Interestingly, such testing also shows a mild abnor-

Schematic diagram of the abnormal reflex mechanism in the developmental of algodystrophy. ¹Initial afferent sensitising spinal WDR neurones; ²Subsequent afferent stimulating sensitised WDR neurones. DRG=dorsal root ganglion; SG=sympathetic ganglion; WDR=wide range multireceptive neurones.

malinity in the normal hand of these patients and is in keeping with some bilaterality of the spinal cord abnormality as suggested previously.^{11 20}

Osteoporosis on x ray examination (patchy in early or diffuse in late cases) is a useful finding but it is not present in many cases. Isotope bone scanning is as useful and probably superior in early cases.²¹ Three phase scans using technetium-99m methylene diphosphonate recording initial (blood flow), early (blood pool), and delayed (tissue uptake as an indicator of active turnover) uptake are advocated as increased vascularity may be the only abnormality, especially in subjects with early or mild disease. Reduced rather than increased uptake is commonly the abnormality seen in children and adolescents, however.²²

There are no consistent abnormalities on routine blood testing, but diabetes mellitus, hyperthyroidism, hyperparathyroidism, and type IV hyperlipidaemia are predisposing factors.^{2 21} Increased urinary hydroxyproline excretion may be found in early disease.²

Management must be aimed at the restoration of movement and function to desensitise the abnormal reflex, and so physical therapy is the cornerstone of treatment.²³ Potentially effective alone, adequate participation is often prevented by the severity of pain despite the use of analgesics. The use of various drugs, such as nifedipine, propranolol, phenoxybenzamine, or even high dose oral corticosteroids, may produce benefit in some patients. Transcutaneous nerve stimulation may be helpful, especially in children,^{24 25} and parenteral calcitonin can be useful,²⁶ especially in early ankle and foot disease. Suppression of sympathetic hyperactivity is logically best achieved by sympathetic blockade, but invasive specialist techniques are needed, often requiring multiple blocks, and may still fail to produce prolonged benefit.^{23 27} An alternative to stellate ganglion or paralumbar blockade for peripheral lesions is a simpler Bier's block technique using intravenous guanethidine.²⁸ This form of guanethidine is not available in the United States and has led to the use of other substances, including reserpine, ketanserin, and local steroids plus lignocaine, which are less effective or have more side effects, or both. Prolonged blockade can also be achieved by infusions of local anaesthetic through an epidural catheter and shows promise, often in association with the use of epidural opiates.²⁹ Surgical sympathectomy has been used in selected cases. To be most effective an active rehabilitation programme used in conjunction with these techniques is likely to be needed.

Although algodystrophy may be a mild transient phenomenon requiring little more than encouraging use of the affected part, treatment of the well established case is more difficult. Treatment within one year of onset is most successful,³⁰ presumably before reflex patterns, tissue changes and any psychological response to chronic pain become fixed. Although often self limiting, many cases persist for years and may become permanent.

Greater understanding of central neurological and local tissue mechanisms is needed. Interest-

ingly, at both sites neuropeptides may have a role. Sympathetic nerve fibres containing vasoactive intestinal polypeptide have been found in abundance in bone.³¹ Vasoactive intestinal polypeptide promotes bone resorption and reduces bone blood flow and so might play a part in the pathogenesis of algodystrophy. A study has been undertaken comparing regional and systemic venous concentrations of vasoactive intestinal polypeptide in lower limb algodystrophy, but locally increased concentrations were not found.³² This may suggest that only a large local increase in vasoactive intestinal polypeptide would overflow into the circulation and it is more likely that any excess in 'chronic' algodystrophy is rapidly degraded in the tissues. Assays of the vasoactive intestinal polypeptide content of affected skin and bone are indicated.

Algodystrophy is not a new condition, but management is hampered by lack of any comparative data between different therapeutic regimens. Trials comparing the effectiveness of various treatments carried out prospectively on adequate numbers of patients and incorporating objective clinical assessments are clearly required. Such approaches are to be encouraged if improvements in the understanding of algodystrophy and its management are to be achieved.

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